

Cognitive rehabilitation in non-communicative brain-damaged patients

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Summary

Conscious patients with severe motor and speech disorders have great difficulty interacting with the environment and communicating with other people. Several augmentative communication devices are now available to exploit these patients' expressive potential, but their use often demands considerable cognitive effort. Non-communicative patients with severe brain lesions may have, in addition, specific cognitive deficits that hinder the efficient use of augmentative communication methods. Some neuropsychological batteries are now available for testing these patients. On the basis of such cognitive assessments, cognitive rehabilitation training can now be applied, but we underline that this training must be tailored to single patients in order to allow them to communicate autonomously and efficiently.

KEY WORDS: augmentative communication, cognitive rehabilitation, eye-tracker, neuropsychological assessment, non-communicative patients

Introduction

Patients with severe brain lesions are often left with motor disabilities that impact heavily on their quality of life (QoL). The locked-in syndrome (LIS) provides the most striking example of the limitations that loss of productive language and of voluntary body movements places on an individual's ability to interact with the environment. Patients affected by the "classical" form of LIS (1) are conscious but completely paralysed and can perform only vertical eye movements and/or blinking, hence they depend entirely on their caregivers for their personal care and social activities, and even to communicate their needs or desires. Yet, LIS patients have been

found to express a strong will to participate and to get the best they can out of their life (2).

In recent years several techniques have been devised to increase communicative possibilities in LIS patients (3) and, in general, in conscious subjects with severe speech and motor disabilities, e.g. in amyotrophic lateral sclerosis (4). When patients show remnants of voluntary motion, as in "incomplete" LIS (1), these techniques usually exploit residual movements, but when only vertical eye movements are spared, as in "classical" LIS (1), communication systems usually rely on eye-tracking methods, although other creative solutions have also been devised to overcome communication problems (e.g., 5,6). Thanks to the most impressive technological advances, it is now possible to derive brain signals without resorting to physical movements, through so-called brain-computer interface (BCI) systems (7,8).

It is very important to stress, however, that even the most sophisticated methods and technologies are reserved for patients without cognitive impairments, since lengthy and effortful training is often required in order to use these devices efficiently (10). Patients with severe motor limitations associated with specific cognitive deficits are almost automatically excluded from using communication aids, and this unfortunate association of deficits is probably more frequent in clinical practice than one might imagine. For instance, lesions restricted to the anterior brainstem, causing LIS due to the selective but complete damage of descending motor pathways, are classically considered to have no cognitive consequences (1). However, a survey showed that 14% of LIS patients self-reported attentional problems and 19% reported memory problems (2). Recent investigations of specific cognitive domains have demonstrated that the complete de-efferentation in these patients can cause selective defects, for instance in motor imagery (11,12) or even in processing of facial emotional expressions (13). Most importantly, when LIS patients are extensively assessed using specifically adapted test batteries (see below), several cognitive deficits can be identified; this is particularly true when the brainstem damage is associated with other cortical or subcortical brain lesions (14-16), as is often the case after a traumatic brain injury.

Clearly, therefore, systematic assessment of cognitive abilities is the only means of establishing whether non-communicative brain-damaged patients are likely to benefit from the use of augmentative communication systems, and possibly which device is best suited to the single patient. Cognitive assessment is, of course, severely hampered by the presence of motor and speech disorders and, as a consequence, neuropsychological deficits often go unnoticed in affected patients, but the optimal use of communication aids depends on correct

evaluation of an individual's neuropsychological profile. Moreover, for some cognitive deficits several rehabilitation approaches are now available, which can significantly improve outcome and QoL in brain-damaged patients (17-20). We wish to stress here that such rehabilitation approaches can be adapted for non-communicative patients. From this perspective, the assessment of cognitive functions may represent the starting point for the possible development of specific cognitive rehabilitation treatments, aimed at allowing patients to use communication aids.

In the present paper we briefly describe available augmentative and alternative communication devices and the main instruments for neuropsychological evaluation in non-communicative patients. In the last part of the paper we summarise two pilot case studies, with the intention of illustrating briefly how specific cognitive evaluation can drive rehabilitation aimed at achieving efficient use of communication systems and, ultimately, at enhancing the QoL of non-communicative patients.

Augmentative communication systems

Augmentative and alternative communication (AAC) is the term used to refer to the various modalities used to meet the expressive communication needs of people with significant speech disabilities. The term "augmentative" refers to the possibility of improving the patient's communicative abilities, while "alternative" refers to the use of alternative modes of communication (21). AAC methods have been extensively applied in brain-damaged patients (22) and range from non-technological methods (gestures, signs) to analogical communication tablets and complex computerised systems with dedicated software. Technological advances have led to the development of special input devices, such as modified keyboards and computer mice controlled by small head movements, and computerised eye-tracking systems to record patients' responses (these can also be used to control vocal synthesisers) (10). In his lifetime, an individual may be recommended several AAC systems to optimise his communication and achieve the greatest possible QoL, but attainment of this goal is dependent on the patient's ability to express exactly what he wants to say in the way he wants to say it (23). Achievement of the most effective level of communication depends on several elements, such as the patient's degree of disability, correct evaluation of his neuropsychological profile, and the selection of an appropriate method, tailored to his residual abilities. The main factors leading to abandonment of AAC are related to lack of support and training, and more generally to a poor match between the person and the technology (24). Indeed, in many cases the approach to AAC system selection is unsystematic or idiosyncratic. The AAC assessment process must integrate information about a patient's personal history, clinical conditions, and familiar and social environment in a data-driven and patient-centred manner (10). This strongly suggests that the cognitive profile of non-communicative brain-damaged patients should be carefully evaluated before identifying, selecting, and implementing AAC technology solutions (21).

Cognitive evaluation in non-communicative brain-damaged patients

Non-communicative patients cannot use verbal or motor response modes, and thus the assessment of cognitive deficits has to rely on elementary codes for communication, usually based on eye movements (e.g., looking up for 'yes' or closing the eyes for 'no'). It is important to appreciate that the eye-movement response mode is associated with marked fatigue effects, but nonetheless it allows a formal neuropsychological assessment, as has been shown in a few studies on LIS patients (14-16). Here we summarise some of the methods used in those studies.

Orientation in time and place

Orientation in time and place can be assessed through oral presentation of questions, followed by patient's selection (by eye-coded yes/no response) of the correct answer among orally or visually presented choices (15).

Attention

The capacity to maintain focused attention during a relatively long period of time can be assessed through an auditory attention task, similar to tasks included in computerised test batteries (e.g., 25). For instance, a continuous sequence containing low-frequency, medium-frequency or high-frequency sine wave tones can be presented and the subject has to respond via pre-arranged eye-movements each time he/she detects two consecutive identical sounds (16).

Short-term memory

Standard forward and backward span tasks can be adapted for a yes-no recognition response mode, to assess the capacity to maintain (forward span) and manipulate (backward span) verbal information during a short period of time. Digit (or word) sequences can be orally presented and the patient asked to recognise the corresponding items in a pre-ordered list (16). Delayed-matching-to-sample methods can also be used to assess short-term retention of visual and verbal material (15).

Long-term memory

The capacity to retain and retrieve novel information can be assessed with repeated presentation of verbal or non-verbal material (e.g., lists of words or pictures); patients can then be required to make yes/no recognition judgements for the stimuli presented in the learning phase (distinguishing them from distracters) (15,16).

Language competences

Many tasks can be devised to assess the different aspects of language processing. Oral comprehension can be tested through questions about personal identity or by presenting simple statements requiring yes/no responses (15). Phonological and semantic competences may be assessed by means of auditory-visual picture matching tasks: for instance, patients can be instructed

to produce pre-arranged eye movements only when the picture being shown matches the word (or the sentence) pronounced by the examiner. By using distracters that are phonologically or semantically related to the target pictures it is possible to check different aspects of auditory language processing. The same word-picture matching procedures have also been used to investigate verbal intelligence (by means of adapted versions of picture vocabulary tests) (16), the ability to identify incongruous sentences, and comprehension of implicit information (15).

Logical-mathematical reasoning

Mental calculation and simple problem-solving abilities may be assessed by requiring yes/no selection of correct answers among other choices presented orally or visually (15).

Executive functioning

The capacity to develop and adapt cognitive strategies to a new situation where previous responses are no longer valid can be assessed by adapting executive tests, such as the Wisconsin Card Sorting Test, to yes/no response modes (16).

Visual perception and visuospatial abilities

Visuospatial abilities can be systematically assessed by matching-to-sample procedures. In the following section, an example of assessment of visual exploration abilities is described, in which usual methods to detect unilateral spatial neglect were administered by a computerised eye-tracking system (24).

Cognitive rehabilitation: two pilot cases

To demonstrate the need for specific, tailored cognitive rehabilitation training in conscious non-communicative patients with severe brain lesions, we here summarise the rehabilitation approach adopted in two pilot cases (26,27). In both patients quadriplegia and anarthria were associated with specific cognitive disturbances that hampered the use of a computerised eye-tracking system for communicative purposes. In each case a specific cognitive training programme was devised by applying principles and established methods of cognitive rehabilitation, the aim being to allow these patients to achieve greater levels of interaction with the environment.

Case 1

Case 1 was a 27-year-old right-handed male graduate who sustained a severe closed head injury in a road accident, with multiple micro-haemorrhagic lesions in cortical and subcortical areas of both hemispheres and in the brainstem (26).

The patient was in a coma for approximately one month post-onset, and was then in a vegetative state for 11 months. About one year after onset, the patient began to recover reproducible ocular movements towards acoustic or visual stimuli; sixteen months after onset he

was able to direct his head and gaze, albeit for only brief periods of time, towards people calling him by name or towards people entering his room, but he still presented spastic quadriplegia and anarthria. From that time on, the patient managed to use an eye-code communication system, opening his eyes wide to indicate affirmative responses and shutting them to express negative responses. The patient had relatively spared language comprehension abilities but was densely amnesic: he could recognise his close relatives (parents and brothers) but not friends; he could not recall relatives' ages or occupations and could not answer questions about everyday events or his past history. As far as episodic memory could be tested by means of the eye-code system, retrograde amnesia for at least the past five years and profound anterograde amnesia were detected. Eighteen months after onset, the patient performed conjugated eye movements in all directions and could make small lateral movements of his head, but he breathed through a tracheostomy tube, was dysphagic, amimic, quadriplegic, anarthric, and still densely amnesic; he also had episodes of pathological laughing and crying. Since the eye-code system did not allow the patient to communicate his feelings or needs, we decided to start a rehabilitation training to enhance his communication skills through the use of an eye-tracking system. Because of the concomitant memory deficits, we had to adopt an errorless learning procedure that has been successfully applied in global amnesic patients (17,18).

Rehabilitation training. As a first step, we verified whether the patient could interact with a computerised infra-red eye-tracking system (MyTobii, Tobii Technology, Danderyd, Sweden). In several sessions, on separate days, we asked the patient to pursue a slowly-moving coloured circle on the monitor with his eyes and to fixate it whenever it stopped and flashed. On the first day, the patient's attempts to follow and fixate the target were very poor, meaning that he could not use the system successfully. On the following days, however, the patient's performance improved and from the fifth session onwards the patient performed the calibration procedure with only occasional errors, and could use the infra-red eye-tracking system to answer yes/no questions. However, after six sessions he still denied ever having used or even seen the monitor with the infra-red devices, or ever having met the examiner.

The patient's steady improvement in his ability to use the eye-tracking system (despite his dense anterograde amnesia) prompted us to plan a rehabilitation programme with two main aims: i) to teach the patient to use a simple eye-commanded writing software (based on an enlarged keyboard shown on the monitor); and ii) to render him autonomous in using the eye-tracking system, i.e. in launching and shutting down the desired software, and in switching among available computer programs (a writing software, an augmentative communication software with symbols for basic needs and desires, or simple games based on visual perceptual matching). To achieve these goals, a two-month training program was planned, based on three 40-minute sessions per week. Each session included: i) writing tasks (in which the patient had to fixate the target letters on the screen), aimed at stimulating the patient's ability to manipulate phonological and graphemic representations, to scan the mon-

itor systematically, and to accomplish prolonged tasks; ii) specific training in using a PC via the eye-tracking system. The patient had already used a PC in his premorbid life for simple leisure activities, but when prompted by specific questions, he could not recall any relevant information. The rehabilitation training aimed to teach sequential commands for efficient use of the PC: after repeated demonstration of the commands, the patient was required to repeat the same steps by fixating the appropriate boxes on the monitor. In the recall phase, we strictly followed an errorless procedure, by which errors in patients' responses are minimised (17,18).

Results. At the end of the training the patient's performance improved on most writing tasks, but he never succeeded in writing a single word correctly upon dictation. However, a relevant change in error type was observed: at the baseline most errors were perseverations (50%), unrelated responses (30%) and graphemic errors (20%), whereas at the end of the training most errors were graphemic in nature (omissions, insertions, transpositions or substitutions of letters).

Most importantly, the patient had become autonomous in using the eye-tracker to launch the system spontaneously, to navigate through directories, to find the desired software, and to shift between different programs. Formalised assessments revealed that the patient became fully efficient after a few sessions and progressively required less time and effort to use the system. However, at the end of the training the patient still denied being able to use, or even having ever seen, an eye-tracking system, and also did not recognise the psychologist who had assisted him during the rehabilitation programme.

Case 2

Case 2 was a 57-year-old right-handed housewife who developed multiple ischaemic lesions secondary to endovascular embolisation of a ruptured aneurysm of the right middle cerebral artery. One month after surgery, brain MRI showed lesions in the ventral pons and in the deep periventricular white matter of the right temporoparietal region (27).

After surgery, the patient remained in a coma for about 20 days, and eventually evolved to a vegetative state. Two months after onset she showed reproducible horizontal and vertical eye movements towards acoustic or visual stimuli, and four months later she had gradually learned to use an eye-code communication system, with eyes closed indicating affirmative responses, and had become able to respond to simple verbal and written questions using such a system. At that time, she breathed through a tracheostomy tube and presented amimia, dysphagia, anarthria and spastic quadriplegia. A simple confrontation procedure did not reveal visual field defects, but during formal assessment and in daily life the patient tended to gaze preferentially towards her right side and showed apparent difficulties and delay in moving her eyes towards her left side, a finding compatible with the presence of left neglect in association with incomplete LIS. Since our patient could communicate only by means of the eye-response code, we decided to use a computerised device based on eye-tracking technology to exploit her functional interactive skills.

Assessment and rehabilitation training for neglect. As a first step towards the use of augmentative communication devices, we assessed the patient's ability to interact with a computerised eye-tracking system (MyTobii P1750, Tobii Technology, Danderyd, Sweden), and used the same apparatus to quantify her apparent rightward bias in eye movements. Several visual pursuit and exploration tasks, repeated several times a few days apart, demonstrated that the patient could comply with task instructions and was potentially able to interact with the system but was affected by a strong visual exploration bias towards the right side. The same pattern was observed in two tasks devised to assess eye movements quantitatively, with number of fixations and total observation time showing a significant asymmetry between the two visual hemifields. One month later, virtually the same results were obtained, without significant differences in eye movement parameters between the two assessments. This stable asymmetry of visual exploration precluded efficient use of augmentative communication software, and induced us to devise a specific rehabilitation training program.

On the basis of well-established recommendations for rehabilitation treatment of visuo-spatial neglect (17,18), we implemented a traditional visual scanning training method, administered via the computerised eye-tracking system. Progressive visual scanning exercises were devised, during which the patient was trained to search and fixate the targets, also using verbal encouragement in the event of failure; when the patient achieved a 90% correct response rate on three exercises of a given complexity she was presented with exercises of the next level of complexity. The training, which involved four 40-minute sessions per week, lasted four weeks.

Results. The patient gradually recovered her visual exploration skills during the training. At the end of the treatment she could pursue and fixate targets for most of the time at the most complex level of exercises, thus demonstrating a task-specific improvement. Moreover, re-assessment on the same tasks used at baseline demonstrated an obvious improvement in the patient's ability to scan the visual stimulus arrays, and a significant increase in number of fixations and observation time on the left side. However, the most important result, for our purposes, was the evidence that the patient could start using the augmentative communication software associated with the eye-tracking system.

Concluding remarks

As clearly stated in the introduction, our main aim was to underline that a specific cognitive assessment is necessary in non-communicative brain-damaged patients, in whom neuropsychological deficits, which can hamper the use of communication aids, often go unnoticed. The two pilot studies summarised here provided clear examples of how standard, well-established rehabilitation approaches can be adapted for non-communicative patients. We hope that the present paper might further stimulate the assessment of cognitive functions and the subsequent development of cognitive rehabilitation treatments in non-communicative patients, thereby enhancing their quality of life.

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